
Chapter IV

SOME UNCOMMON CONDITIONS

- ONYCHOGRYPOSIS
- PALMOPLANTAR KERATODERMA
- CALCIUM PYROPHOSPHATE DIHYDRATE (CPPD) DEPOSITION DISEASE
- HYPERKERATOTIC ECZEMA
- NECROBIOSIS LIPOIDICA
- SQUAMOUS CELL CARCINOMA
- DERMATOFIBROSARCOMA PROTUBERANS

ONYCHOGRYPOSIS

A 75-year-old male patient with type 2 diabetes diagnosed at the age of 64 years was referred to the foot clinic for foot care. He was a psychiatric patient treated on an out-patient basis. The patient had findings of peripheral neuropathy with loss of sensation of pain, light touch, vibration and temperature. Peripheral pulses were palpable. Claw toes and extreme onychogryposis was noted (Figure 4.1). His nails were cut using a special nail trimmer. Instruction in foot care was given; extra depth shoes were provided in order to accommodate the deformity. He visited the clinic on a monthly basis and had his nails cut without any other foot problems.

Onychogryposis is caused by chronic repetitive trauma particularly to the nails on the great toe. The nails may be grossly thickened, hard and very elongated (Figure 4.2 shows this condition in another patient). They may be elevated from the nail bed, curved inwards or turned sideways.

The deformed nail can press against another toe causing ulcerations. When the patient does not wear shoes, the deformed toenail often grows vertically. When socks or shoes are being worn, the deformed toenails tend to develop in such a way as to accommodate the clothing.

Keywords: Onychogryposis

PALMOPLANTAR KERATODERMA

A 64-year-old male patient with type 2 diabetes diagnosed at the age of 55 years attended the foot clinic for foot care and instruction in the management of his condition, palmoplantar keratoderma. On examination diffuse thickening of the palmar and plantar skin, together with hyperkeratosis was noted (Figure 4.3). Nail deformities were also observed. He had findings of peripheral neuropathy, while the peripheral arteries were palpable.



Figure 4.1 Onychogryposis



Figure 4.2 Onychogryphosis



Figure 4.3 Palmoplantar keratoderma

The patient was instructed in appropriate foot care. Local debridement with keratolytics was prescribed. Protection from friction with soft insoles may be helpful in this condition.

Palmoplantar keratoderma is an autosomal-dominant trait characterized by diffuse,

thickened hyperkeratosis of the palms of the hands and soles of the feet. The hyperkeratosis may be so thick that the skin may crack, especially in dry, cold weather. Infection with *Tinea pedis* frequently occurs as the fissures provide a portal of entry for the fungus. The nails on

the hands and toes may be dystrophic and become infected with fungus.

Keywords: Palmoplantar keratoderma

CALCIUM PYROPHOSPHATE DIHYDRATE (CPPD) DEPOSITION DISEASE

A 74-year-old female with type 2 diabetes diagnosed at the age of 68 years and treated with sulfonylurea with acceptable diabetes control, was referred to the outpatient diabetic foot clinic for possible osteomyelitis of her fifth left toe. She had intense pain at this site when resting and walking. The pain started after the patient had worn a tight pair of shoes for a few hours.

On examination, redness, edema, and callus formation were noted at the outer aspect of the left fifth toe (Figure 4.4). She had findings of diabetic neuropathy (no

sensation of vibration, no Achilles tendon reflexes, but she could feel pinpricks; vibration perception threshold was 45 V on both feet). Peripheral pulses were palpable.

Debridement of the callus revealed a cheesy material emanating from the base of a superficial ulcer. A culture of this material did not reveal any microorganisms. A plain radiograph showed radiodense deposits at the articular bursae of the distal interphalangeal joint; no osteomyelitis was apparent (Figure 4.5). Examination of this material with compensated polarized light microscopy showed rhomboid-shaped and weakly positive birefringent crystals, which is typical of CPPD deposition disease.

The patient was advised to rest. She visited the foot clinic on a weekly basis for callus debridement. The ulcer healed completely in 3 weeks.

CPPD deposition disease (or pseudogout) of the foot joints may pose a problem with diagnosis when the location is atypical. The knee is the most frequent joint affected by pseudo-gout, followed by the



Figure 4.4 Painful inflammatory lesion of the fifth toe, due to calcium pyrophosphate dihydrate deposition disease



Figure 4.5 Radio-dense deposits at the articular bursae of the distal interphalangeal joint of the fifth toe due to calcium pyrophosphate dihydrate deposition disease

wrist, shoulder, ankle, elbow and hands, although every joint can be affected. Treatment includes rest, aspiration of the joint fluids, and systemic use of non-steroidal anti-inflammatory medication.

Keywords: Pseudo-gout; calcium pyrophosphate dihydrate deposition disease; CPPD deposition disease

HYPERKERATOTIC ECZEMA

A 65-year-old male patient with longstanding diabetes visited the outpatient diabetic foot clinic for a chronic pruritic lesion of his left foot.

On examination, he had severe diabetic neuropathy and peripheral pulses were palpable. A hyperkeratotic lesion with dense yellowish scales over a red skin patch was observed on the plantaro-lateral aspect of his left foot (Figure 4.6). The scales were firmly adherent on the epidermis, and not

easily debrided. Dry skin on the heel was also present.

The patient was referred to the dermatology department for treatment.

This situation occurs on the palms of the hands and soles of the feet, almost exclusively in men. It may result from irritation or allergy, although the cause is usually unknown. Topical moisturizers containing lactic acid or urea are applied after soaking the affected area for 20 min. Topical coal tar preparations may be applied daily under occlusion if severe lichenification is present. Per os antihistaminic medication, or low-dose corticosteroids may be of some help for short periods.

Keywords: Hyperkeratotic eczema

NECROBIOSIS LIPOIDICA

A 50-year-old lady with type 1 diabetes diagnosed at the age of 38 years, visited the outpatient clinic for control of her



Figure 4.6 Hyperkeratotic eczema. Hyperkeratotic lesion with dense yellowish scales over a red skin patch. The scales are firmly adhered to the epidermis and not easily debrided with a blade. Dry skin on the heel

diabetes (HbA_{1c} : 7.0–7.9%) on a regular basis. She was free of micro- or macro-angiopathy. Two years after her diabetes was diagnosed, the patient noticed a few small, red, irregular, violaceous papules on the dorsum of her feet. These papules slowly enlarged, coalesced and became scaly, irregular plaques, with minimal central atrophy, and an advancing red border (Figure 4.7). New lesions appeared on her left ankle and right leg. Apart from causing cosmetic problems, the lesions were asymptomatic.

A biopsy of these lesions showed necrobiosis lipidica (formerly ‘necrobiosis lipoidica diabetorum’), an unusual disorder of unknown mechanism, strongly associated with diabetes mellitus but also found in subjects with normal or abnormal glucose tolerance.

Typically, such lesions occur on the anterior shin of both lower legs, but they also may be located on the arms, hands or head. They may precede the diagnosis of diabetes, and sometimes they



Figure 4.7 Necrobiosis lipidica. Scaly, irregular plaques, with minimal central atrophy and an advancing red border

are pruritic, dysesthetic or painful. They ulcerate—usually after a trauma (shown in Figures 4.8 and 4.9 in other patients)—in approximately 35% of the cases, but do not usually lead to infection.

Histological examination of the lesions shows necrobiosis, which provides the foci for ‘hyalinized’ collagen bundles (Figure 4.10), fibrosis, histiocyte infiltration (Figure 4.11) and granulomata (Figure 4.12). Capillary walls become thickened (Figure 4.10).

Topical application of corticosteroids may have fair results against progression of



Figure 4.8 Ulcerated lesions associated with necrobiosis lipoidica. Note a not ulcerated lesion over the epiphysis of the fibula



Figure 4.9 Infected ulcerated lesions associated with necrobiosis lipoidica

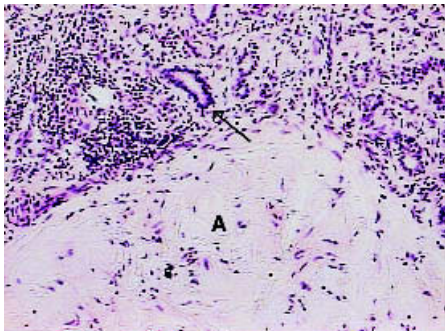


Figure 4.10 Histological findings in samples taken from the patient with necrobiosis lipoidica shown in Figure 4.7. A, foci of ‘hyalinized’ collagen bundles (necrobiosis), fibrosis, and histiocyte infiltration. Thickened capillary walls are evident (arrow). H&E stain, ×100

the lesions. Various other agents have been tried, such as aspirin or pentoxifyllin, with mixed results.

Keywords: Necrobiosis lipoidica

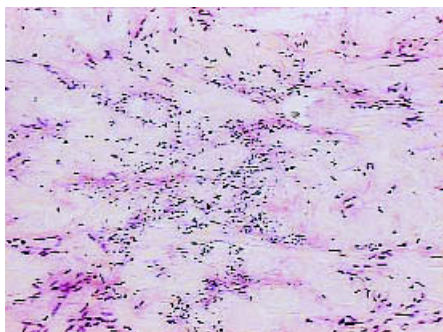


Figure 4.11 Histological findings in samples taken from the patient with necrobiosis lipoidica shown in [Figure 4.7](#). Foci of 'hyalinized' collagen bundles (necrobiosis), fibrosis and histiocyte infiltration can be seen. H&E stain, ×100

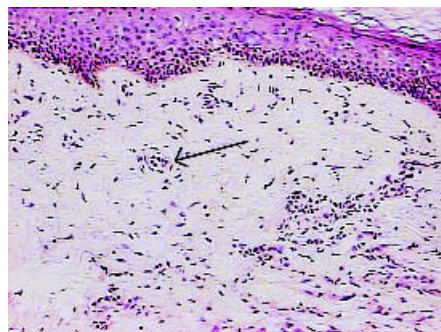


Figure 4.12 Histological findings in samples taken from the patient with necrobiosis lipoidica shown in [Figure 4.7](#). Note the presence of a granuloma (arrow). H&E stain, ×100

SQUAMOUS CELL CARCINOMA

Squamous cell carcinoma (SCC) developed on a neglected burn scar in a 48-year-old diabetic male patient. SCC is the second most common skin cancer after basal cell carcinoma. It arises from the dermis and it is most common in areas exposed to the sun. It is an aggressive and invasive

cancer; it may penetrate underlying tissues, and it metastasizes in distant tissues, lymph nodes, and organs. Presentations vary and for this reason the neoplasm is difficult to diagnose. Pink, red or tan plaques, ulcers ([Figure 4.13](#)) or erosions and scaling may be apparent. Secondary SCC arises in areas of old scars, especially burn scars, chronic non-healing wounds and radiation lesions (Marjolin's ulcers). Plastic surgeons used a free latissimus dorsi musculocutaneous flap to repair the defect, after extensive



Figure 4.13 Squamous cell carcinoma presented as an ulcer on the lateral aspect of the foot of a diabetic patient. The design of the excision and the recipient vessels are indicated. (Courtesy of O. Papadopoulos)



Figure 4.14 Free latissimus dorsi musculocutaneous flap used to repair the defect, after extensive excision of a squamous cell carcinoma. Patient whose foot is shown in [Figure 4.13](#). (Courtesy of O. Papadopoulos)

removal of the cancer, within a healthy border ([Figure 4.14](#)).

Keywords: Squamous cell carcinoma

**DERMATOFIBROSARCOMA
PROTUBERANS**

Recurrent dermatofibrosarcoma protuberans (DFSP) was diagnosed in a 71-year-old

male diabetic patient. DFSP is an uncommon aggressive soft tissue sarcoma of low malignant potential, arising in the dermis of young to middle-aged adults and it is slightly more frequent in men than women (57 versus 43%). It is most commonly located on the trunk and proximal extremities. Initially it presents as an asymptomatic bluish, red or flesh-colored nodule with a diameter of a few millimeters



Figure 4.16 A free latissimus dorsi musculocutaneous flap was used to repair the defect, after extensive excision of a dermatofibrosarcoma protuberans of heel. Patient of [Figure 4.15](#). (Courtesy of O. Papadopoulos)



Figure 4.15 Ulceration of recurrent dermatofibrosarcoma protuberans of the heel of a diabetic patient. The design of the excision and the recipient vessels are indicated. (Courtesy of O. Papadopoulos)

to >20 cm located on top of plaque-like lesions, or as superficial ulceration of some of these nodules. It infiltrates the surrounding tissues and, if untreated, it may ulcerate (Figure 4.15). It may recur after surgical excision and lead to metastases. Moh's micrographic surgery, using wide margin resection, is the mainstay of treatment.

Plastic surgeons used a free latissimus dorsi musculocutaneous flap to repair the defect, after wide removal of the cancer (Figure 4.16).

Keywords: Dermatofibrosarcoma protuberans